ASIG

AUSTRALIAN SCLERODERMA INTEREST GROUP

SCLERODERMA connections

PATIENT NEWS

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greetings,

The original motivation for establishing the Australian Scleroderma Screening Program was to provide a service to people with scleroderma and their doctors, for screening for the serious complication of pulmonary arterial hypertension (PAH).

But why all the fuss about PAH?

Timely identification of PAH can be complex, particularly in a busy rheumatology practice. The "one stop shop" visit to a screening centre for tests and clinical assessment aims to take the worry out of this process. While an elevated pulmonary arterial pressure on an echocardiogram is a strong predictor of PAH, this often occurs late in the disease, so detection relies on a combination of other factors including reduced diffusing capacity on lung function tests and reduced exercise tolerance. Unfortunately, people with scleroderma can have a poor exercise capacity for so many reasons, making it difficult to unravel the cause of their symptoms. The primary goal of the screening program is to increase rates of detection of PAH in its early stages. In addition, the spin-off Australian Scleroderma Cohort study is driving multiple research studies, some of which aim to find better markers of PAH in its early stages and which are summarised elsewhere in this newsletter.

These are worthy objectives indeed, but in all the complexities of screening tests and data collection, we must not lose sight of the individual caught in the vortex of all this activity, who is being poked and prodded, while all the while hoping that a right heart catheter (required for diagnosis of PAH) is never recommended and if it is, that PAH is never detected. Undoubtedly, this can be a serious and debilitating complication and a source of much anxiety but it is not all bad news. De-mystifying some of the issues around PAH is one of the important components of managing this disease.

Firstly, 90% of patients with scleroderma will never develop PAH. It is only because better non-invasive methods for early detection are yet to be developed, that international guidelines currently recommend annual screening. Newer therapies have measurable benefits in this condition. Furthermore, it appears likely that earlier detection and treatment leads to improved survival so there is much to be optimistic about. While there has been a growing and rewarding collegiality between the various specialists who treat this disease – cardiologists, respiratory physicians and rheumatologists – the "glue" which holds the PAH clinic together is the PAH nurse. In the patient version of this issue, Leah McWilliams, PAH nurse at the Royal Adelaide Hospital, the centre featured in this issue, provides helpful advice for the new patient with PAH. The clinical support and pastoral care provided to people with PAH and scleroderma by these dedicated PAH nurses is without doubt, second to none.

Susanna

Susanna Proudman
ASIG Chair

donations.

ASIG Research Program

Thank you to the families and friends of the late Mrs Rebecca Haddock and Mrs Brenda Matear for their generous donations to the research fund. Donations assist a number of our projects such as blood sample research, maintenance of the national database and funding of a PhD student.

Anyone wishing to make a donation to the fund should contact the Project Co-ordinator, Jill Byron on jill.byron@svhm.org.au or phone (03) 9288 3986.

World Scleroderma Day

This day occurs in June. It is a good opportunity to educate health professionals and the wider community about scleroderma.

This year, at St Vincent's Hospital the scleroderma team held an information session for staff across the hospital. Presentations were given by Dr Wendy Stevens and Barbara Gemmell focusing on the clinical aspects of patient care, and the fellows Dr Vivek Thakkar and Dr Owen Moore gave an overview of their research projects. It was well attended and included nurses from the wards and from in-home care, technicians from the respiratory clinic and others interested in learning about the disease.

The lunch sponsored by Actelion was much appreciated.

A/Prof Susanna Proudman and Leah McWilliams

Royal Adelaide Hospital

Leah McWilliams began training as a nurse at the Royal Adelaide Hospital in 1987. She writes, "On completing my training, I secured a permanent position as a registered nurse in an orthopaedic/ rheumatology ward and soon became a clinical nurse. Travel beckoned, resulting in twelve months of backpacking in America, Europe and South Africa. On returning to Adelaide, I completed a Bachelor of Nursing degree at Flinders University and found myself back at the RAH in the Orthopaedic/ Rheumatology Outpatient department. It was here I was approached by A/ Prof Susanna Proudman to take up the position as clinical studies manager for the Rheumatology Unit.

I embarked upon a very steep learning curve, managing a clinical trial in patients with early rheumatoid arthritis and another in pulmonary arterial hypertension. I was also enlisted by the cardiology and respiratory physicians to assist with Medicare applications for highly specialised drugs for PAH. The staff weren't quite sure what a rheumatology nurse was doing in their OPD, but were welcoming, interested and accommodating.

The scleroderma screening program grew fast and we now have over155 patients enrolled, so my feet didn't really hit the ground till a new cardiac nurse came on board early this year.

My main role is to be a first point of contact for patients, answer questions, provide educational support, liaise with pharmacists, GP's, specialist physicians, community nurses and research staff. I prepare and collate patient files to help streamline patient visits, perform joint examinations and complete lots of paperwork! I provide support to the clerical staff by making sure appointments are as convenient as possible for patients. Often I catch up with patients admitted to the wards, but the aim is to provide enough 'accessible' support to keep admissions to a minimum.

It is A/Prof Susanna Proudman's everyday example that is a privilege to be near and the quiet bravery of our patients that makes working at the RAH so extraordinary.

A career highlight was receiving a 'Commendation award' on Susanna's nomination for services to 'Safety & Quality' within the South Australian Health Service; oh and having a patient name his race horse 'Leah' after me!"

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Often in clinics in the mornings, best time to catch me is afternoons in the office

So you have been told you have pulmonary arterial hypertension – what now?

By Leah McWilliams

Having attended screening examinations as part of the Australian Scleroderma Screening Program (ASSP), you are now faced with a diagnosis of pulmonary arterial hypertension (PAH).

What now?

Well thankfully, as a participant in the ASSP, the diagnosis of PAH will more than likely have been made in a speedy manner, giving you the best opportunity for early treatment and an improved outcome.

With the advent of new and effective therapies, there is good reason to believe the combined care from your rheumatologist, cardiologist & respiratory physician will give you the very best chance for improvement.

Treatment for PAH usually involves one of a range of new "PAH-specific" medications which must be approved by Medicare Australia. An application is sent to Medicare Australia on your behalf. The results of your right heart catheter, echocardiogram and six minute walk test must be included in the application to demonstrate your eligibility for government subsidised treatment. The new medication is classified as a "Highly Specialised Drug" and can only be prescribed by your specialist in a hospital and not by your GP. The echocardiogram and 6 minute walk test must be repeated every six months so that applications for further scripts can be made. If you do not have the tests required for re-application in a timely manner, further scripts cannot be approved so it is important that you always attend your appointments

Tips whilst taking a "Highly Specialised Drug"

- Don't lose the Medicare approved script! - Always keep it in a safe place, or leave it on file with your pharmacist.
- Your pharmacist will not have this medication in stock, so remember to always lodge your script with your pharmacist at least one week prior to you requiring it, as delivery to your pharmacy may take up to 5 days.
- If you are admitted to any hospital, you must take your own supply of medication with you and importantly, take it home on discharge.
- Whilst on the PAH specific medication, it is very important to have a blood test every month. The results will go to your GP & prescribing physician.
- Never stop medication abruptly.
- Ensure you always have enough medication with you whilst on a holiday.
- "Red flags" for seeking help from your doctor:
  - chest pain, increasing shortness of breath or lack of energy, fainting, rapid palpitations, ankle swelling
- Remember, if you have any queries, your PAH/scleroderma nurse is just a phone call away!

ASIG research output

With longitudinal projects it can be several years before it is possible to analyse in a meaningful way the data that are collected. ASIG will be presenting several abstracts at the second World Scleroderma Congress to be held in Spain next February.

Our PhD fellow, Dr Vivek Thakkar, submitted three abstracts on behalf of the group.

As many of you are aware, one of the key initiatives set up by ASIG was a screening program designed to identify scleroderma patients who may be suffering from the serious complication of pulmonary arterial hypertension, or 'high blood pressures in the lungs'. This enables patients to be identified at the earliest possible time with commencement of therapies that improve quality of life. This is an exciting area of research in Australia and abroad as evidence is emerging confirming programs such as ours improve the quality of life and life expectancy of patients. We are committed to finding better ways of screening for this problem, and have undertaken research to see if simple blood tests can be used to help us identify patients at highest and lowest risk for this condition. We have also been studying a number of markers that may offer insights into the driving forces leading to the development of pulmonary arterial hypertension in scleroderma. One such blood test, called N-Terminal Pro Brain Natriuretic Peptide has shown particular promise with preliminary results identifying not only patients who have the condition, but also suggesting others who should be closely monitored and reassured, and a third group who seem to be a very low risk of developing PAH. This is an exciting area of research that we hope to develop over the next twelve months and incorporate into our screening program. Needless to say this work is only possible because of the many patients who have kindly donated their blood to further scleroderma research.

Dr Jane Zochling's abstract entitled "An immunochip based interrogation of scleroderma susceptibility variants" has also been accepted.

St Vincent's Rheumatology Fellow, Dr Owen Moore, has been working on an ILD project with ASIG. He submitted two abstracts on behalf of the group. He has provided a summary of his research and initial findings:

It is recognised that interstitial lung disease can be a major problem and cause of death in some patients with scleroderma. We sought to test the ability of a simple scoring system for the amount of lung disease on CT chest scanning to predict the outcome of patients with scleroderma lung disease. We were able to do this thanks to the large amount of data collected within the ASIG database, the support of local centres in examination of clinical notes and the kind assistance of enrolled patients in sending us their lung scans. Three experienced doctors rated the scans as either showing extensive or limited disease.

Our data showed two useful findings. Firstly, that a finding of extensive lung disease is highly predictive of a poor outcome. Secondly that, while the scan is useful initially, over time the most important test to establish outcome is the pulmonary function test with worsening results over the subsequent follow-up period indicating more serious progressive lung disease.

We hope that these findings will help the treating specialists to target therapy to those who are most likely to need it, and to reassure those who have less severe disease that it is not likely to be a problem for them in the future and to reduce the number of CT scans patients need to undergo.

Additional Research

One manuscript has been submitted to a journal and one is in the final stages of writing. We'll keep you posted on how these go.

Further information about ASIG can be found at:
http://rheumatology.org.au/rheumatologists/asig-public.asp

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